A151 RECEPTORS FOR B CELL ACTIVATING FACTOR OF THE TNF FAMILY (BAFF) ARE EXPRESSED IN MUSCLE TISSUE OF MYOSITIS PATIENTS WITH ANTI-JO-1 OR ANTI-RO 52/ANTI-RO 60 AUTOANTIBODIES AND CORRELATE WITH PLASMACYTOID DENDRITIC CELL MARKERS

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Background A role for B cells in the pathogenesis of myositis is supported by occurrence of autoantibodies. B cell activating factor (BAFF) plays a role in autoantibody production and is inducible by type I interferon (IFN- α/β). Polymyositis patients (PM) with anti-Jo-1 autoantibodies have elevated serum levels of BAFF that correlate to serum creatine kinase levels. The significance of this finding for muscle involvement/function is unclear. Sera of this subset of myositis patients and those with anti-Ro autoantibodies have an IFN- α/β inducing capacity and plasmacytoid dendritic cells (pDCs) as well as IFN α/β inducible myxovirus resistance-1 (MX-1) protein are present in muscle tissue.¹

B lymphocytes and plasma cells (PC) with evidence for local affinity maturation are occasionally found in muscle tissue of myositis patients. B cells can differentiate to plasma blasts upon exposure to IFN- α/β from pDC. Plasma cell survival and antibody secretion is enhanced by BAFF. Upregulation of the BAFF transcript was recently described in muscle tissue of dermatomyositis (DM), PM and inclusion body myositis (IBM) patients.² Whether this is related to autoantibody profile is not known. Here we postulate that this could be of particular

relevance for patients with anti-Jo-1/anti-Ro autoantibodies (anti-Jo-1/anti-RoAbs).

Objective To investigate expression of the receptors for BAFF, its correlation with autoantibody positivity and presence of pDC marker (BDCA-2) and MX-1 in DM, PM and IBM muscle.

Methods Biopsies from 23 patients, (10 with anti-Jo-1/anti-RoAbs) and 7 healthy controls were investigated for expression of BAFF-R, BCMA and TACI (Zymogenetics) and 19 of these also for B cell marker (CD19) and PC marker (CD138). The number of BAFF-R positive cells per area (mm^2) was compared with areas/ mm^2 positive for BDCA-2 and MX-1.

Results B cells or PCs were present in 2/15 or 10/15 patients respectively (2/2 or 5/10 with anti-Jo-1/anti-RoAbs). BAFF-R, TACI and BCMA expression was seen in 5, 7 and 7 of the 23 patients, more frequently in patients with anti-Jo-1/anti-RoAbs compared to patients without these autoantibodies or controls (KW: $p=0.007$, 0.07 and 0.03). BCMA, TACI and CD138 expression correlated with BDCA-2 expression ($r_s=0.56$, 0.43 and 0.7 ; $p<0.05$). Number of TACI⁺ cells correlated with MX-1⁺ areas ($r_s=0.43$; $p<0.05$).

Conclusion Our data support a possible local effect of BAFF, which could lead to autoantibody production in muscle tissue of myositis patients with anti-Jo-1 or anti-Ro52 autoantibodies, and this could be induced by IFN- α/β produced by pDCs. These data indicate a role for BAFF in disease mechanisms in autanibody-positive myositis.

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